Impaired joint mobility in Guillain-Barré syndrome: a primary or a secondary phenomenon?

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Abstract
Three patients with Guillain-Barré syndrome had significant residual impairment of joint mobility. Pain in the limbs and axial skeleton was a prominent early feature, as were autonomic disturbances and bulbar involvement resulting in prolonged mechanical ventilation. All three patients developed marked joint stiffness and contractures despite having physiotherapy from the outset. The skeletal problems and complications became major components of disability despite improving neurological status.

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Guillain-Barré syndrome is best regarded as a clinical syndrome rather than a specific disease entity. Classically the neurological features develop after episodes of viral or other infections and the syndrome is infrequently associated with autoimmune, neoplastic and other diseases. The prognosis in idiopathic cases is generally good, with residual distal weakness as the commonest cause of long term disability. A recent review described persisting moderate or severe motor deficit in 5–30% of cases.[1] Although regular passive movements and splinting to avoid contractures are often recommended, reports of long term follow up do not describe contractures or other joint problems as a persisting cause of disability. We describe three patients with Guillain-Barré syndrome in whom reduction in the range of joint movement contributed significantly to the disability.

Patients and methods
All the cases described fulfilled the criteria for the Guillain-Barré syndrome defined by the National Institute of Neurological and Communicative Disorders and Stroke.[2] Thus all presented with predominantly progressive motor weakness, which was relatively symmetrical, loss of reflexes, mild sensory loss and facial nerve palsy and autonomic dysfunction. The patients were referred from different acute neurological units to a neuro-rehabilitation unit after prolonged periods of care. Improvements in functional ability were assessed using the Barthel Index,[3] which is a measure of self-care and mobility scored on a range from 0 (total dependence) to 20 (independent).

Case 1
A 57 year old salesman was admitted in July 1989 with a history of pain in the extremities, peripheral sensory disturbances and progressive muscular weakness preceded by a week's history of pharyngitis, neck adenitis and conjunctivitis. He had flaccid quadriparesis, areflexia and sensory loss in the extremities to mid forearms and knees. His condition deteriorated rapidly with bulbar involvement and dysautonomia characterised by labile hypertension, urinary retention and faecal incontinence. He was ventilated for a period of 10 weeks. His course was complicated with recurrent chest infections and deep vein thrombosis. The patient complained of persistent and severe axial pain and marked symmetrical joint stiffness affecting most joints particularly the small joints in both hands (figure).

On admission to the rehabilitation unit, eleven months after the acute onset, he was totally dependent on nursing care and was bed bound. He complained of backache and right hip pain as well as pain in both shoulders, elbows and wrists on passive exercises. A skeletal survey and a bone scan showed no evidence of active inflammation although the hip radiograph showed evidence of tendon calcification. The haematological and biochemical screen including LFT and ESR were normal. His autoantibody screen and serology for Lyme disease were negative. His neurological state improved progressively with return of sensations and muscle bulk and the appearance of weak tendon reflexes. His progress, however, was hampered by articular disability. There was some improvement in proximal joint movement range over 12 months: shoulder flexion increased from 70°–120°; abduction from 50°–80°, elbow flexion from 70°–120°; and hip and knee flexion both from 80°–110°. Distal joints, however, showed minimal change, for example, wrist flexion from 45°–55°.

Functionally he regained continence in sphincteric functions, was able to feed himself with adapted cutlery and mobilise independently in an electric wheelchair with an adapted manual control knob, and was able to use a computer and typewriter with a stick held in the mouth. His ability to transfer improved although he still required assistance with this and with bathing and toileting, reflected in an improvement in Barthel score from 0–7.

Case 2
A 54 year old woman was admitted in May

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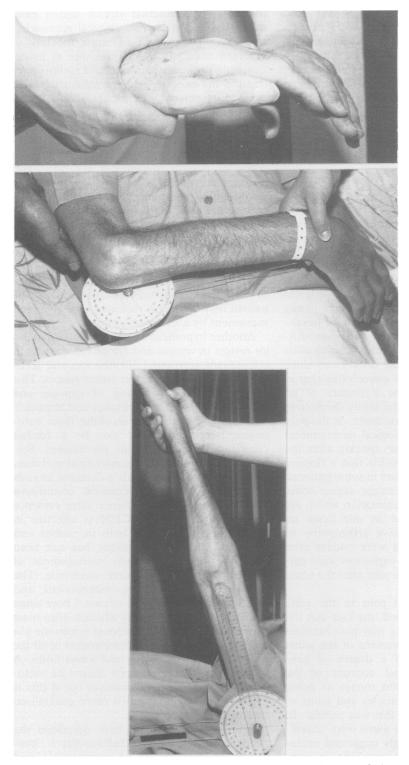B Pentland
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Figure Limitations in passive movements (case 1). Top: Metacarpo-phalangeal flexion limited to 20°. Middle: Elbow flexion limited to 70°. Bottom: Shoulder flexion limited to 70°.

1987 with paraesthesiae in the tongue and gums following an upper respiratory tract infection. Subsequently there was a rapid progressive generalised weakness, dysarthria and dysphagia and she was maintained on mechanical ventilation for five months. Past history included a partial right lower lobectomy for pulmonary tuberculosis at the age of sixteen, ruptured ovarian cyst and hysterectomy at the age of twenty nine and depression for which the patient was maintained on regular imipramine.

In the early stages she exhibited episodes of bradycardia alternating with tachycardia and significant fluctuations in blood pressure. She also suffered two attacks of lobar pneumonia. She experienced severe limb pain requiring control with morphia and was given a trial of oral steroids. Her records indicated that she received physical therapy in the intensive care unit and throughout her stay in hospital. Different radiographs of the hands, hips, pelvis and lumbar spine revealed osteoporotic changes without erosive features or heterotropic calcification.

On admission to the rehabilitation unit she was dysarthric and there was evidence of bilateral lower motor neuron facial nerve palsy. A flaccid quadriparesis was evident worse distally. She was areflexic but had no sensory abnormalities. The initial problem with joint pain had settled to a large extent and the patient was maintained on simple analgesia. The patient was totally dependent on nursing care. There were marked joint contractures and these were treated with serial splinting and passive stretching. Despite satisfactory neurological progress shown by grade 4 power in the shoulders and elbows and grade 3 power at the wrists, she was left with fixed contractures of shoulders to 80° flexion and 40° horizontal abduction as well as contractures of the metacarpo-phalangeal, proximal and distal interphalangeal joints bilaterally. The patient was finally discharged home 32 months after the onset of her condition. Her Barthel score rose from 1 to 6. She was supplied with an electric chair and Possum environmental control and remained dependent on nursing care for help with washing, dressing and feeding.

Case 3

A 70 year old woman was admitted in July 1990 with ascending progressive muscular weakness affecting initially the legs and then the arms over a 24 hour period. She was areflexic with minimal sensory impairment and had rapid bulbar involvement and needed to be ventilated for 4 months. She had plasmapheresis and was fed enterally. The course was complicated with dysrhythmias, hypertension, hypercalcaemia and chronic constipation.

Following discharge from ITU, she complained of marked stiffness and pain in the small joints of the hands, shoulders and wrists bilaterally and this was exacerbated by passive mobilisation of the joints involved. Hip, knee and ankle joints were also stiff with a reduced range of movement. Physiotherapy was administered regularly in ITU. She had intra-articular steroid injections in both shoulder joints with significant improvement noted three days later.

Investigations one year after onset of her condition showed a normal autoantibody screen, ESR 40, slightly raised C3 and C4 levels with normal immune complex ratio. Antibody titres for Lyme disease were insignificant. Radiographs of both hands showed evidence of osteoporosis with tendon sheath calcifications. Goniometric measurements of joint movement showed an improvement in

shoulder abduction from 80° to 90° and extension 25° to 40°, wrist flexion 55° to 75°, limited metacarpophalangeal flexion to 60° and interphalangeal flexion to 20° during 6 weeks of rehabilitation. There was also noticeable return of muscle bulk in the forearms and weak proximal stretch reflexes. Pain in the different joints has settled to a minimum and sensations have recovered fully. The patient is still dependent on nursing care for most activities of daily living but has made progress as an inpatient.

Discussion

Pain is a well recognised accompaniment of the acute phase of Guillain-Barré syndrome occurring in about half of the reported cases.[4-6] This is most frequently back pain[6 7] or buttock and other large muscle discomfort[5 6] but can be prominent distally as in Landry's original description.[5] Explanations of the source of this pain include changes in the muscle of neurogenic origin,[5] poor posturing and joint stiffness, meningeal irritation[8] and inflammation of the cutaneous nerves and distal roots stimulatory C fibres.[8] It is also conceivable that in some cases pain may be a feature of an inflammatory process in the joints themselves and/or the periarticular structures. As the pain often precedes the neurological deterioration and therefore follows more quickly after the infective episode, it is possible that a fleeting post-infective arthritis occurs in some patients. If this was the case one might expect some inflammatory signs on examination which are not usually present. Also in our cases no radiological signs of erosive arthropathy or inflammatory joint disease were evident even though these radiological exposures were carried out approximately one year after the acute onset.

Whatever the cause of pain in the early stages of the cases described, the fact that the joints were painful to move may have resulted in inadequate passive movement of the joints resulting in stiffness and a degree of contractures. In her personal account of the condition Bowes[9] described nurses as being reluctant to stretch her muscles and joints to the point of resistance as this was painful. In our cases the first patient's joints were "fixed" in a position which strongly suggested inadequate passive movements resulting in contractures. The second patient had pains of such intensity that they required narcotic analgesia and this phenomenon has also been described by others. Faced with such, degree of pain, the therapist or the nurse may be understandably cautious about putting the joints through the full range of movement.

Prolonged immobilisation is known to result in poor muscular compliance with structural histopathological changes in muscle and connective tissue. Lack of stretch, when muscles are immobilised in the shortened state, and loss of contractility are the two main contributory factors to such changes.[10] This is followed by joint stiffness, muscular shortening and largely irreversible contractures. It is clear

therefore that muscular stretch and/or electrical stimulation are two physical approaches to prevent contractures. Despite the lack of reported cases of contractures due to inadequate physiotherapy, a number of authors have stressed the need to perform regular repeated passive movements and the use of splints and pillows to maintain the position of function.[11 12] There is, however, perhaps a case for a more precise structured programme determining the frequency and duration with which range of passive movements are done and looking prospectively at the cause of the disorder and subsequent disabilities in patients with severe early joint pains.

In contrast to the supposition that joint immobility results from inadequate physiotherapy, it is possible that inappropriate excessive passive movement could result in capsular tears or other damage to joints. In the presence of hypotonia and sensory impairment the patient may be unable to detect excessive joint movement by a therapist or nurse.

Another hypothesis is that muscular paralysis results in venous and lymphatic stasis with consequent nutritional disturbances and accumulation of tissue fluid in tissue spaces. This will result in a softening of capsule and ligaments followed by adhesions and fibrosis.[13] Vasomotor disturbances resulting from autonomic neuropathy may also be a further contributory factor to this mechanism. Perhaps another method of passive joint mobilisation that is associated with a decrease in such risk, is the use of mechanical continuous passive motion (CPM). There is some evidence showing that intensive CPM is effective in maintaining joint range both in rabbits and humans,[14] but this technique has not been evaluated yet in the early management of patients with Guillain-Barré syndrome. The presence of severe pain, osteoporosis and smooth skin in the fingers of case 3 bore some similarity to algodystrophy which is seen most commonly in the shoulder hand syndrome but this patient had bilateral involvement of all the major joints in the upper and lower limbs. A reflex neovascular dystrophy cannot be excluded as a pathogenic mechanism but if this is the case it must be a much more generalised variant than usual.

Several recent reports have described the residual disability after Guillain-Barré syndrome in populations of patients[1 6 15] and residual weakness is reported to be the major problem with no reference made to loss of or reduced joint mobility. Some reports describing joint problems requiring surgery in children with the condition are available[16 17] but these are mainly concerned with scoliosis and foot drop correcting procedures. Our patients may represent the chance occurrence of atypical cases but all three had profound disability and social handicap over one to three years after acute neurological presentation primarily as a result of reduced joint mobility rather than neurogenic weakness.

One could therefore argue the case for intensive monitoring and periodic measurements of joint range of movement as a standard

part of management from the outset to identify patients at risk of developing articular complications. Perhaps it is also time to review the different aspects of nursing care and physical therapy with specific recommendations towards integrated standardised methods for mobilisation and care of joint structures.

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